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CONGENITAL ANGULATIONS AND FRACTURES OF THE EXTREMITIES

By

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These affections of the skeleton, although morphologically rather simple, present etiologic and therapeutic problems far more complicated than those met with in the ordinary birth fractures—which are acquired fractures most often due to obstetric manoeuvres during delivery (*Watson-Jones,* 1946; *E. T. Madsen,* 1955).

Their *occurrence* is rare. In the obstetric departments of the Copenhagen University Hospital were found 60 fractures of the long bones in 105,119 births (0.056 %) during the period 1920–1949, and less than one tenth of those were true congenital fractures. In 154,119 patients treated in the Orthopoedic Hospital of Copenhagen from 1935 to 1949, only 7 cases were found (0.0045 %). Therefore, it is impossible to form an opinion of etiologic and therapeutic factors based on personal observation of a sufficiently great number of cases, and extensive studies of literature are necessary.

Many various theories have been advanced to explain the *etiology* of congenital fractures and angulations, and it must be admitted that these lesions are "extremely embarrassing when etiology is concerned" (*Freund,* 1936):

Trauma to the pregnant mother's abdomen was formerly considered of prime importance (*Kramer,* 1900; *Rentoul,* 1912). Many such cases have been reported (*Gurlt,* 1857), and they are of certain medico-forensic interest, but most often the infants were inadequately examined —before the roentgen era (*Snure,* 1929) and the cases reported, occurred at a time when knowledge of congenital fractures was poor.

It is very unlikely that a trauma to the mother's abdomen might injure the foetus during the first months of pregnancy, and in the

later months, as a rule, abortion or premature labour is induced. However, a few cases of intra-uterine fractures following a trauma shortly before birth have been reported (*Smith*, 1913; *V. Madsen*, 1953). In their clinical course such cases behave like ordinary birth fractures with rapid healing and restitution of function.

It is questionable whether *v. Büngner*'s 5 cases of fractures of the lower leg (1891) could be attributed only to the obstetrician's traction on the feet of the child in podiac presentation. It is more likely that some pre-existing bone defiency was the real cause of the eventual pseudarthrosis but could not be diagnosed (without x-ray).

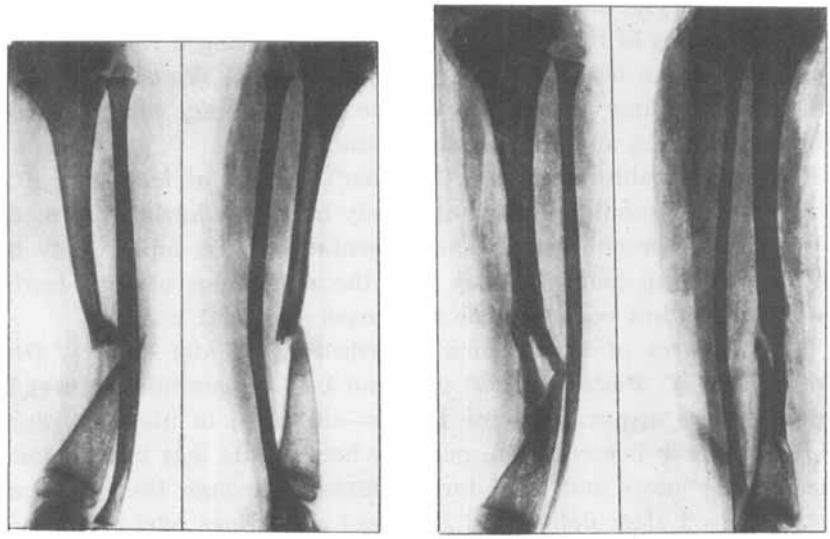
The fractures of the femora described by *Hölder* (1917), *Otten* (1940) and *E. T. Madsen* (1955) were not true congenital fractures but fresh fractures acquired during labour—although in utero—with the child in cephalic presentation, possibly because its legs had remained flexed at the knees and hips during passage through the pelvis, and were fractured after delivery of the head and upper part of the body as the back got stuck against the coccyx. *Murken*'s case (1927) had probably the same mechanism but here a pre-existing skeletal deficiency may be assumed as the child exhibited other congenital anomalies: spina bifida and pes varus.

Mechanical pressure in connection with oligoamnion or hydramnion has been suggested as a probable cause of congenital fractures and other malformations (*Bludau*, 1893).

Browne (1936) in a very interesting paper tried to explain the mechanism. It seems, however, unlikely that abnormal intra-uterine pressure alone would be sufficient to produce fractures if the bones of the foetus are normally developed, and many more cases would then be expected. Moreover, it has been proved by other writers that most congenital fractures appear already during the earlier months of pregnancy. Possibly, intra-uterine pressure may impair an existing deformity.

Amniotic bands or adhesions were formerly often thought to bring about congenital fractures (*Sperling*, 1892) but they were never convincingly demonstrated. *Granzow* (1930) reported a case which in his opinion was due to amniotic bands but nothing abnormal could be observed in the placenta or the membranes. *Ollerenshaw* (1925) does not believe in amniotic band lesions after he found exactly identical malformations in the left leg of two twin sisters. He supports the theory of a developmental defiency.

Strangulation by a too short umbilical cord with fracture of the femur was reported by *Green* (1899) and *Henning* (1900) but in their case pre-existing anomalies in the bone could not be excluded.



a b

Fig. 1 A.

(Case 1, girl B. E. H.) Congenital pseudarthrosis of the left tibia. Angulation was noticed at 3 months of age, fracture occurred at 14 months, and pseudarthrosis was present from 2 years of age. Now seen at 5 years, showing postero-lateral angulation, pointed ends with sclerosis and over-riding.

Fig. 1 B.

The same case, 10 months after an *Albee* grafting operation with a transplant from the right tibia. A fracture line is visible in the transplant.

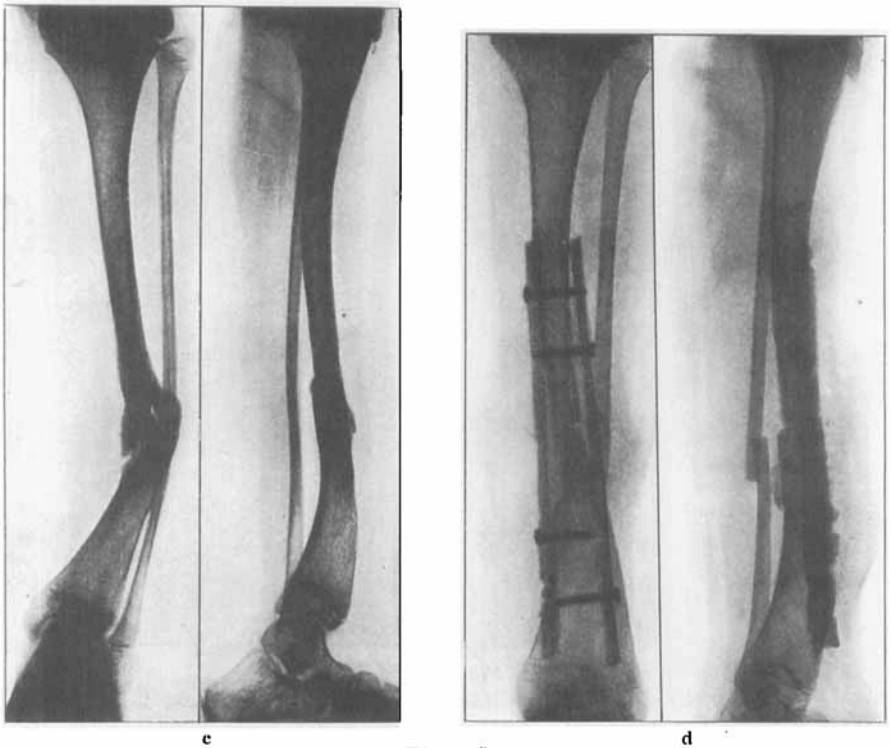
Degeneration and shortening of striated muscle cells was demonstrated in cases of congenital angulation of the tibia by *Middleton* (1934). He claimed that tibial angulations were always anterior and this was ascribed to prenatal lesions of the hamstring muscles with fatty degeneration.

Several cases of posterior angulation of the tibia have, however, been reported (*Heymann & Herndon*, 1949; *Miller*, 1951). The degeneration of muscles found by *Middleton* might well be a secondary or an associated phenomenon.

Vascular anomalies, especially a defective nutrient artery with secondary skeletal dysplasia may theoretically cause angulation or fracture with pseudarthrosis (*Henderson*, 1925; *Boyd*, 1941; *Heymann & Herndon*, 1949). But it is difficult to prove that this lesion is primary.

The same holds true of *disturbances of the nervous system*, particularly the vasomotor nerves (*Bentzon*, 1924; *Moore*, 1949).

Defective ossification may be caused by several factors in addition to the above mentioned:



c d

Fig. 1 C.

There remains considerable lateral angulation and pseudarthrosis 4 years after the grafting operation.

Fig. 1 D.

Another grafting operation has been performed 6 years after the first one. Two full-grafts from the mother have been fixed to the fragments with vitallium screws and with bone chips from the patient's iliac crest. The fibula has been divided. Solid healing at 15 years of age, but pes calcaneus and 4 cm shortening.

Inflammation was formerly considered of great importance, especially congenital syphilis. Deformities of the long bones are sometimes seen due to syphilitic osteochondritis with epiphyseal separation and hyperplastic periostitis (*Frølich*, 1941) but these lesions are very different from the true congenital fractures and angulations. In *Camurati*'s review of 145 cases of pseudarthrosis (1930) syphilis never occurred.

Staphylococcus osteomyelitis is occasionally seen in the newborn (*Abesser*, 1953; *Rivier & Vulliemin*, 1953), and may bring about deformities of the long bones, but they are not likely to be mistaken for true congenital fractures. *Freudenberg, Roulet & Nicole* (1952) reported a single case with deformity of the upper femoral epiphysis and

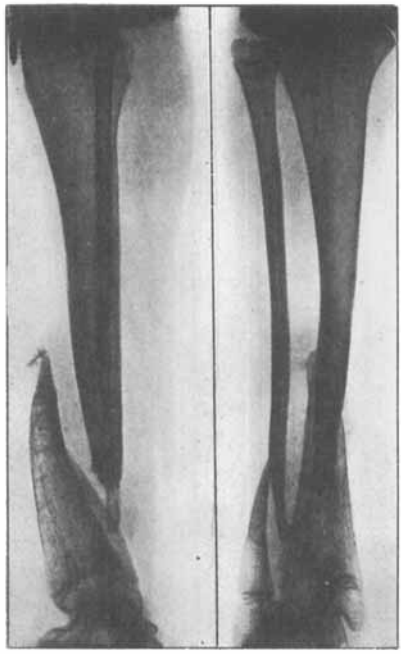


Fig. 2 A.

(Case 4, boy J. L. N.) Congenital pseudarthrosis of the right lower leg. Anterior angulation noticed at 14 months, fracture occurred at 5 years, subsequent pseudarthrosis. Now seen at 12 years of age, with anterior angulation at the distal third of the tibia and the fibula, pointed ends, marked over-riding and 7 cm shortening.

congenital hip dislocation which was ascribed to congenital infection with coxackie virus.

Rickets has been demonstrated in connection with fracture of the fibula in a newborn child of a mother with osteomalacia (*Hauser*, 1953) but this is a very rare condition in the Western countries. *Müller & Sissons* (1951) have reported a case of renal rickets with angulation of the lower leg resembling congenital angulation or fracture sequelæ.

Infantile scurvy and *coeliac disease* may occasionally produce infractions or pathologic fractures; in the latter disease, union of the fractures may be remarkably slow unless vigorously treated (*Watson-Jones*, 1946).

Alltogether, avitaminosis cannot be considered to play any direct rôle in the etiology of congenital fractures.

Hyperthyreoidism and *osseous tumours* as the cause of fractures are exceedingly rare in infants, and the same holds true of reticulosis. A case with infraction of the tibia and the fibula in a newborn, due to eosinophilic granuloma was reported by *Garsche* (1953).

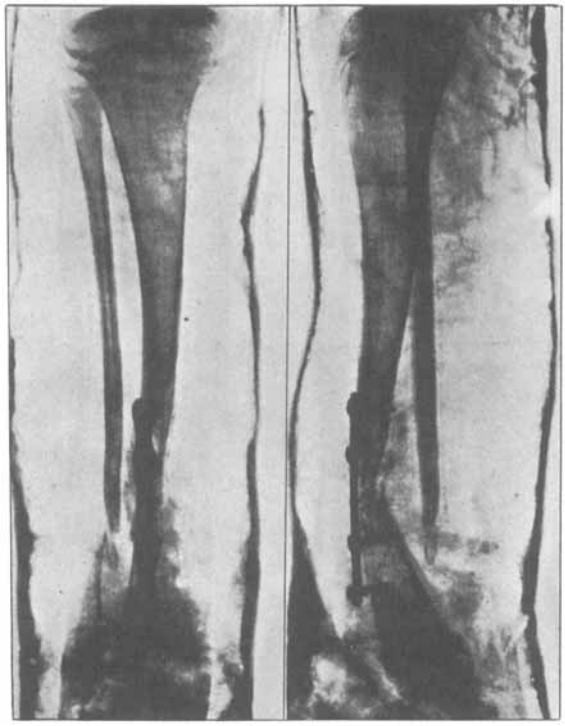


Fig. 2 B.
The same case after operation 2 months later in which was used a vitallium plate and four screws. Already after 6 weeks anterior angulation was again noticed and there was no intention of healing.

A definite connection between *v. Recklinghausen's neurofibromatosis* and congenital pseudarthrosis has been noticed for some years. *Ducroquet* (1937) reported 10 cases with congenital fractures, "café-au-lait" spots and, sometimes, cutaneous neuromas; some of them were hereditary. *Nørgaard* (1937) found that scoliosis very often accompanied neurofibromatosis, and he also mentioned osteomalacic lesions with spontaneous fractures as well as periosteal cysts and lengthening or shortening of the extremities. *Moore* (1941) reported 3 cases of neurofibromatosis with congenital pseudarthrosis. He suggested that neurofibromatosis produces an endoarteriitis which influences the ossification processes. *Miller* (1953) described the characteristic skeletal disturbances in neurofibromatosis with the following three manifestations: localized osseous neurofibromatosis, complex malformations, and general osteomalacia of the *Milkman* type.

Compére (1936) found a relationship between *v. Recklinghausen's localized osteitis fibrosa* and congenital pseudarthrosis. *Russel* &

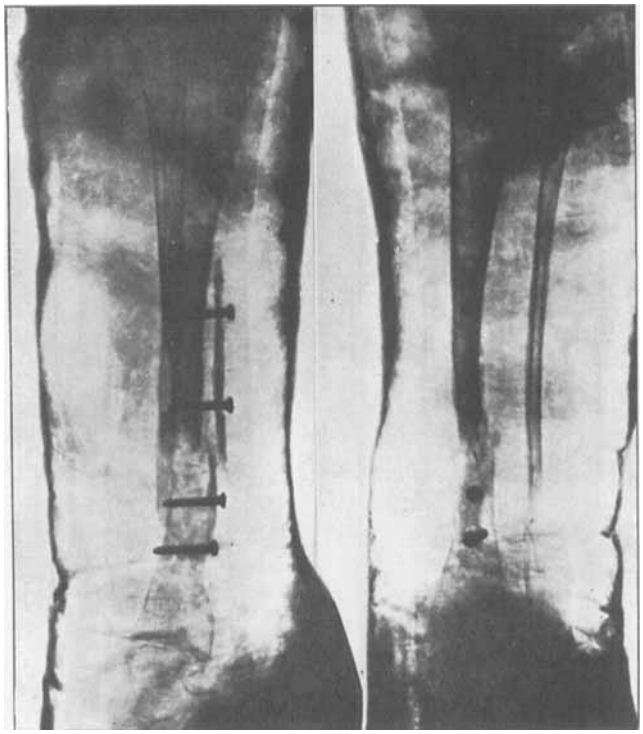


Fig. 2 C.
The vitallium plate was removed 4 months after the previous operation, and a full-graft from the left tibia was placed on the anterior surface of the pseudarthrotic tibia and fixed by four screws with bone chips between the bone ends.

Chandler (1950) employ the term *fibrous dysplasia of bone* which includes monostotic and polyostotic lesions—with replacement of cortical and cancellous bone and marrow by fibrous connective tissue—and the *Albright syndrome*; in the latter is seen skeletal disturbances, pubertas præcox and pigmentation. *Valls, Polak & Schajowics* (1950) have made thorough studies of the pathological anatomy of these dysplasias.

Both neurofibromatosis and osteitis fibrosa may exist simultaneously in the same patient (*Clark, Lamberton & Mathews*, 1953) and are possibly related (*Thannhauser*, 1944; *Aegerter*, 1950). *McCarroll* (1950) presumed that all manifestations of neurofibromatosis arise from a primary developmental defect.

Congenital fractures and *osteogenesis imperfecta* are closely associated. In the more severe form, osteogenesis imperfecta congenita (Vrolik), the children are, as a rule, still-born or die within the first hours of birth. There are found multiple fractures and deficient de-

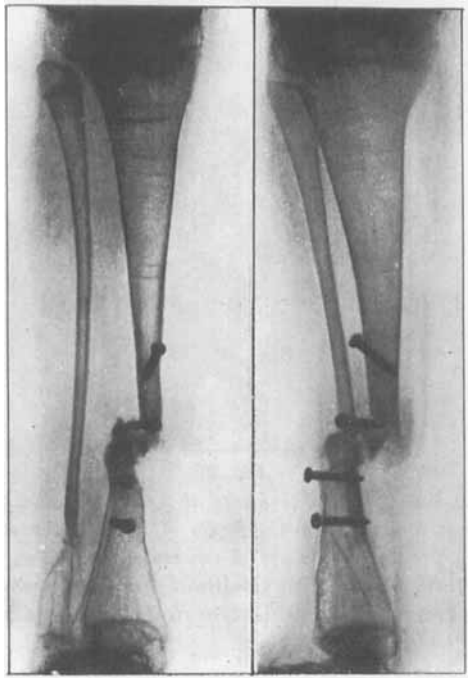


Fig. 2 D.

The result has been completely unsuccessful, there is fracture of the graft, bayonet-shaped axis, marked pes calcaneus and 8 cm shortening. The bone structure is definitely pathologie with a very thin cortex and transverse lines through the diaphysis. The bone ends are sclerotic, the medullary canal is closed, and there is no healing. Amputation was performed 2½ years later.

velopment of the osseous system with caput membranaceum and micromelia (*Seedorff,* 1949). Osteogenesis imperfecta tarda or osteopsatyrosis idiopathica (Lobstein) is a hereditary disturbance of ossification (*Pedersen,* 1940; *Seedorff,* 1949) combined with blue sclerotics, hyperlaxability of the joints, and late deafness (*Fraser,* 1934–35; *Harnett,* 1934–35). Some writers believe that it is an endocrinous disorder, whereas others suggest a congenital defective development of the mesenchyma.

Osteosclerosis fragilis (Albers-Schönberg) also shows a strong tendency to fractures (*Nielsen,* 1942). The long bones are of uniform structure, "marble bones", without roentgen-visible spongiosa. This disease is exceedingly rare, and its origin is unknown. *Watson-Jones* (1946) has pointed out that the bones which show tendency to fragility are more like chalk, whereas those resembling marble are solid.

In 1952 *Duraiswami* succeeded in elucidating the hitherto obscure etiology of congenital fractures and angulations through an excellent

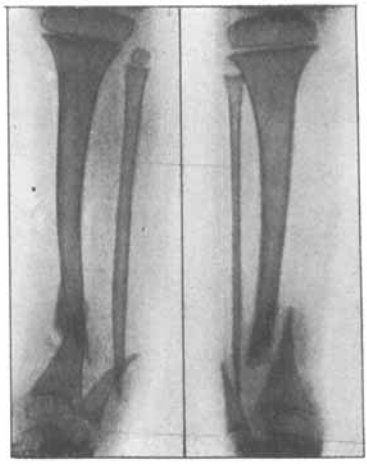


Fig. 3.
(Case 5, girl C. I. H.) Fracture at 1½ years of age with subsequent pseudarthrosis of the right lower leg, now seen at 4½ years of age. Postero-lateral angulation in the lower third with pes calcaneus and 4 cm shortening. Two unsuccessful operations have been performed, one with vitallium plate and screws, the other with a graft from the left tibia. The final result unknown.

experimental work on chicken embryos. By injecting insulin or other agents in various doses into fecundated hens' eggs at various days after incubation he was able to produce skeletal deformities ranging from localized angulations or fractures to generalized bone defects as osteogenesis imperfecta. The fractures, sometimes, were associated with other anomalies such as congenital club foot, absence of the fibula or other bones, congenital dislocation of the hips, and spina bifida with myelo-meningocele, just as is often encountered in the human.

He suggests several different agents which may injure the normal bone development *during the early weeks of foetal life* in human pathology: faulty diet of the mother with defiency of calcium, phosphorus, iodine, iron or vitamins A, B, C, or D; irradiation injuries from x-rays or radium; infectious diseases as rubeola, chicken-pox, measles, mumps; metabolic disorders as diabetes and hypoglycemia; hormonal anomalies; diseases or mechanical disproportion in the uterus; lesion of the placenta; toxemia during pregnancy; ectopic pregnancy; intra-uterine anoxia or asphyxia; and iso-immunization (Rhesus-types). *Duraiswami* furthermore succeeded in transferring the experimentally produced skeletal defects to second and third generation of the animals; in other words, these malformations had become *hereditary*.

It appears conclusive that the majority of intra-uterine fractures, congenital pseudarthroses and angulations are malformations caused

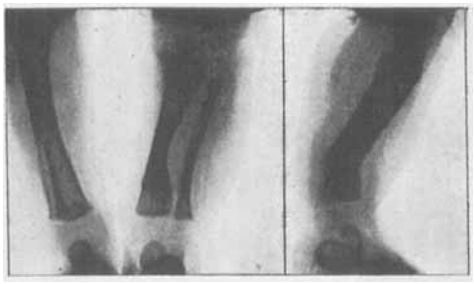


Fig. 4 A.
(Case 7, boy P. E. A.) At birth a fracture of the left lower leg was noticed, the foot being completely dorsi-flexed up along the lower leg. The fracture is still visible 3 months after manipulative reduction and treatment in plaster cast.

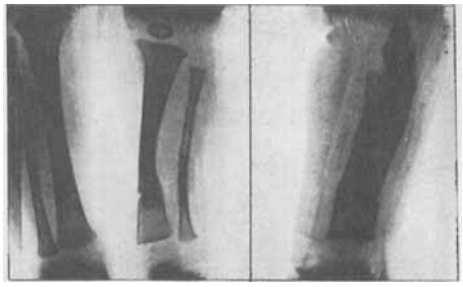


Fig. 4 B.
After a second attempt of reduction the alignment is satisfactory.

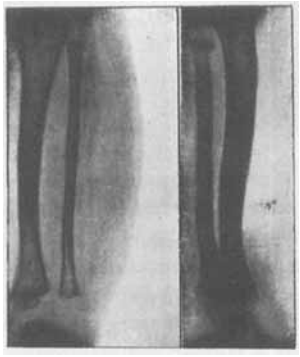


Fig. 4 C.
After one year, healing is complete. It has not been necessary to operate. At 13 years of age the patient walks well with only 1½ cm shortening.

either by genetic, hereditary factors or by some noxious agent to the maternal organism or the foetus during the first 3 to 6 weeks of pregnancy.

Probably v. Recklinghausen's neurofibromatosis and fibrous dys-

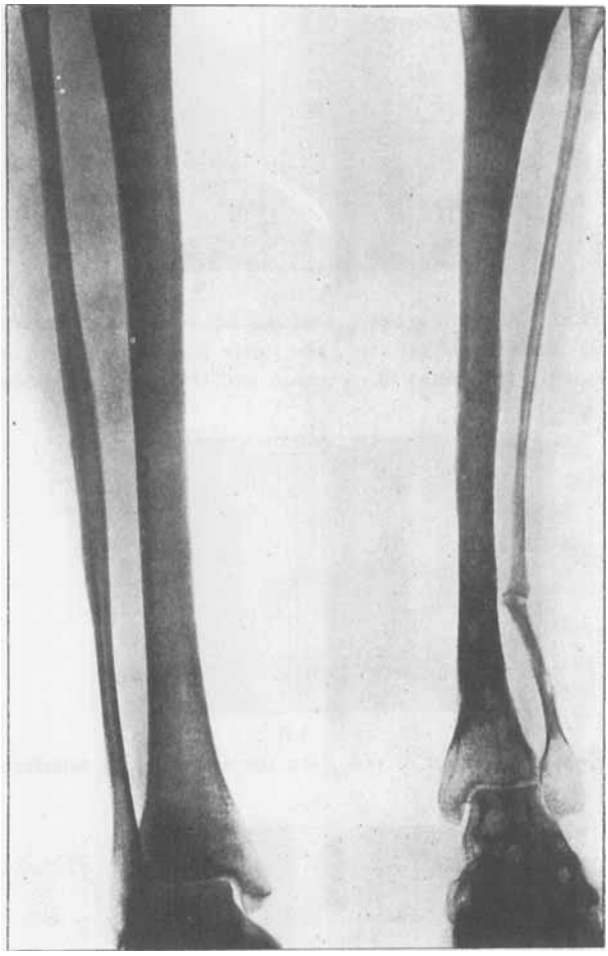


Fig. 5.
(Case 8, girl I. R. R.) Anterior angulation of the left lower leg noticed at 8 months of age. Localized osteitis fibrosa with fracture at 14 months treated by osteotomy. One year later pseudarthrosis, operated by bone grafting with a 10 cm long transplant of *os purum*. The result was unsuccessful, re-operation 3 years later. The proximal end was inserted into the medullary cavity of the distal end. Thereafter healing was obtained, but after 11 months the patient had another fracture at the same site, which was treated by a fibular peg inserted in the medullary canal of the tibial fragments. After 14 months pseudarthrosis again occurred. *Beck*'s drilling was performed, and the patient had a caliper for 8 years. There remains, at 18 years of age, a considerable anterior angulation and a few cysts, but the tibia is solid.

plasia as well as osteopsatyrosis with blue scleræ are different manifestations of congenital disturbance of the mesenchyma. Ectodermal derangement may have taken place too (*Uhlmann & Grossmann*, 1940; *Aegerter*, 1950). Chondrodystrophia foetalis (*Trier-Mørch*, 1941), dys-

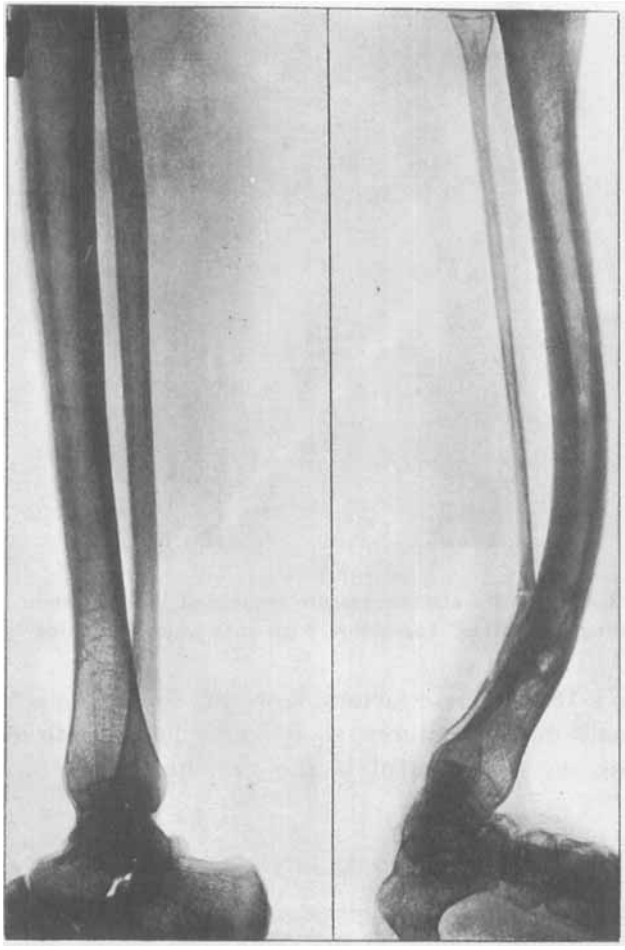


Fig. 5.

chondroplasia (*Bentzon,* 1924; *Hunter & Wiles,* 1934–35), chondrodysplasia calcificans congenita (*Schönenberg & Shallock,* 1953) and artrogryposis multiplex congenita (*Lewin,* 1925) may be of similar etiology.

A defective ossification involving a large area of the diaphysis would explain why congenital fractures heal so badly or recur so often, whereas traumatic fractures in infancy show such remarkably rapid healing and even fractures due to localized tumours unite solidly as soon as the tumour has been surgically removed.

Some congenital fractures, for instance those associated with osteogenesis imperfecta, produce normal callus, but there is a great tendency to re-fracture. Therefore, mechanical factors as unsuitable weight-bearing transmission at the fracture site and inadequate immobilisation may also be of importance (*Hallock,* 1938; *Watson-Jones*

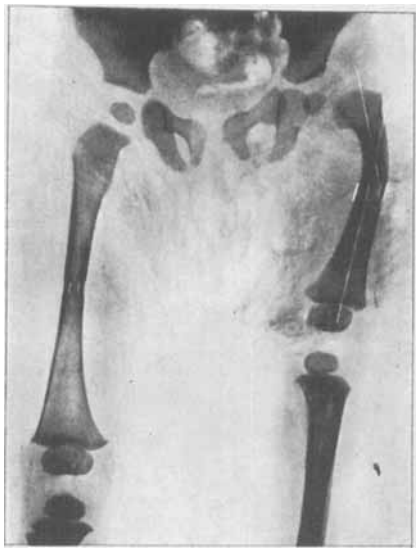


Fig. 6.
(Case 9, girl A. E. H.) Intra-uterine fracture sequelæ of the left femur. Considerable shortening and lateral angulation with coxa vara. The bone is solid.

1946; *Wilson,* 1941). These factors, however, do not impair the results after traumatic birth fractures, so it seems justified to assume that defective osseous development is the principal factor in congenital fractures.

SYMPTOMS AND SIGNS

Angulation, fracture and pseudarthrosis are various forms, or sometimes transitory stages, of the same bone lesion. Most often the tibia is involved, unilateral or bilateral, and not infrequently both the tibia and the fibula are affected. Next often follows the femur, and here the lesion is quite often bilateral. Lesions of the forearm, the humerus and the clavicle are very rare (*Moore,* 1949).

The lower leg.

In the slight cases, only *angulation* is seen, with maximal bending between the lower and middle third of the tibia. Occasionally it is located at the upper third (*Camurati,* 1930). In the majority of cases there is *forward angulation* with talipes equinus, probably due to the pull of the powerful hamstring muscles. All of *Camurati*'s 27 cases showed forward bending or fracture with forward angulation. Antero-medial angulation was more frequent than antero-lateral or directly anterior angulation.

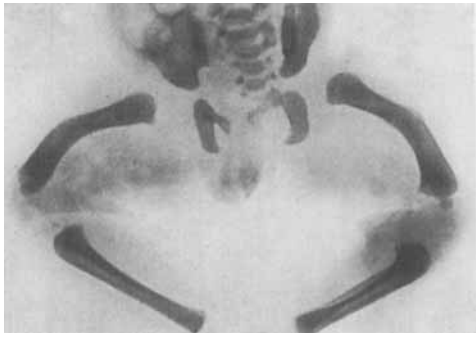


Fig. 7 A.
(Case 10, girl A. M. H.) Intra-uterine fracture sequelæ of both femora with lateral angulation. The medullary canals are interrupted at the middle of the shaft.

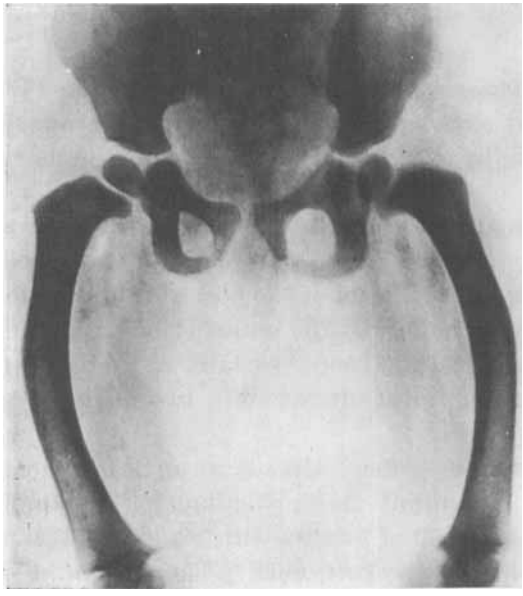


Fig.7 B.
The same case after 2 years. Angulation is less pronounced, the medullary canals are now normal. No operative treatment. The child walks well.

Sometimes a dimple can be seen in the skin at the top of the convexity. It was formerly thought to be a scar from a perforating bone fragment (*Kramer,* 1900) or a pressure ulcer (*Browne,* 1936) but *Middleton* (1934) claims that dimples contain no scar tissue and are only depressions in the skin due to a thin subcutaneous layer over the maximal flexure of the bone.

Cases of *backward angulation* with talipes calcaneus have been

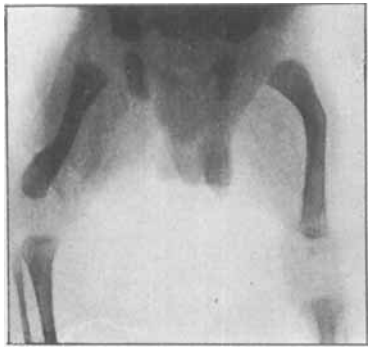


Fig. 8.
(Case 11, boy P. V. N.) Intra-uterine fracture sequelæ of both femora, with lateral angulation at the upper third of the left femur, antero-medial angulation at the lower third of the right femur. The patient had blue sclerotics and one year later his right lower leg was fractured. Probably osteogenesis imperfecta.

reported by *Dawson* (1949), *Heymann & Herndon* (1949) and *Miller* (1951). In eight out of nine cases it was a postero-medial angulation. The tendo Achillis was lax, and usually a dimple could be found posteriorly.

As a rule, angulation is not very marked at first, and the tibia is only moderately curved. The leg is somewhat shortened, and the child walks with a slight limp. Most of the posterior and some of the anterior angulations remain solid, but the majority of the latter only represent a latent stage at birth and sooner or later a fracture occurs at the site of maximal bending, most often within two years of age, following a trivial trauma.

A *fracture* may be present already at birth but most often it is not observed until the infant starts standing or walking. Probably the special walking position of infants with tendency to lean forwards may exert a shearing strain (*Scott*, 1939). The congenital fracture shows the common symptoms and signs of ordinary fractures of the extremities, but it has a strong tendency towards formation of pseudarthrosis in spite of thorough treatment.

The *pseudarthrotic* tibia is considerably shortened, the fracture is completely loose, and the distal parts or the whole leg become atrophied. The angle between the fragments becomes increasingly acute, the foot gets out of its normal position, and there is tightening of the tendo Achillis. Movements in the hip and the knee are usually not affected.

Apart from the above mentioned malpositions of the foot other anomalies are occasionally observed, especially congenital club feet,

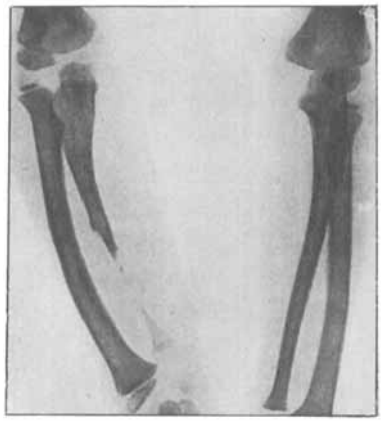


Fig. 9 A.
(Case 13, boy J. G. H.) Neurofibromatosis of the right ulna and lateral angulation of the radius as seen at 6 years of age. The angulation had been noticed when the patient was 2 years old. No treatment.

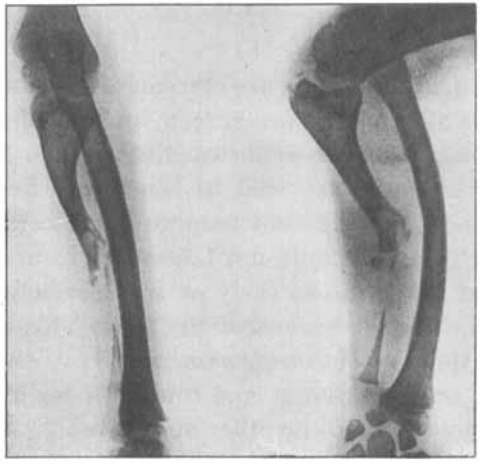


Fig. 9 B.
The same case after 4 years. Increased deformity and distinct bone cysts at the middle of the ulna. Dislocation of the radius at the elbow.

absence or hypoplasia of the fibula, absent metatarsals or toes, and dislocation of the hip or the knee (*Freund,* 1936). Kyphoscoliosis is not uncommon.

The femur.

The femur when affected is considerably bent already at birth, as a rule in the upper third of the shaft but sometimes in the lower third. There may be very marked shortening and a tendency to dislocation

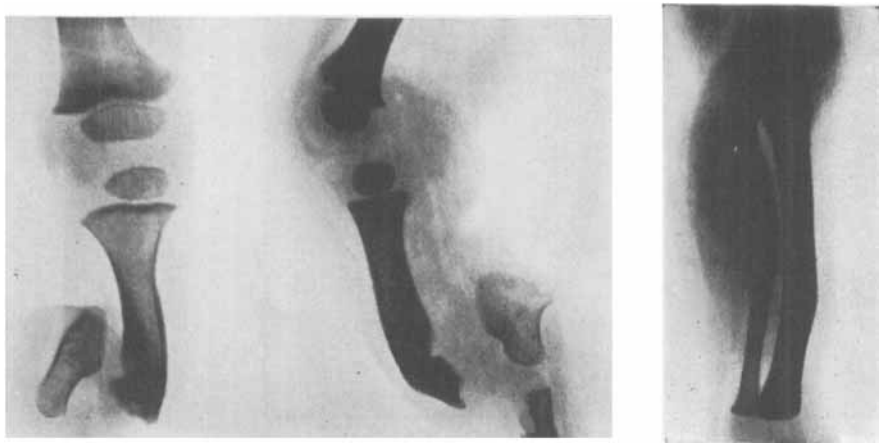


Fig. 10.
(Case 16, boy P. H.) Multiple malformations. Anterior angulation of the left lower leg with solid looking bones. 90° anterior angulation of the right tibia in the lower third. The right fibula and the lateral toes are absent. Slight lateral angulation at the middle of the right femur.

of the hip. Limping and scoliosis are extreme if the lesion is not treated. Only in rare cases a fresh fracture is seen, and congenital pseudarthrosis of the femur has not been reported. Contrary to *Middleton*'s view, angulations of the femur may well be considered healed intrauterine fractures in a bone which has not been so badly injured during early foetal life that solid union could not take place in utero; probably the noxious agent did not work so early or so massively in these lesions as was the case in similar lesions of the tibia. This would be in conformity with the theories of *Duraiswami.*

In some cases both the femur and the lower leg are involved (*Lee,* 1934–35), and there may also be other anomalies.

The forearm.

Lee (1934–35) has described a case of congenital absence of the lower two thirds of the ulna and bending of the radius which was dislocated posteriorly so that its upper 2–3 cm were placed behind the humerus. In our own material are two cases, one with hypoplasia of the ulna and angulation of the radius, the other with bilateral deformities of the forearm and malformations of the hands.

Diagnosis.

Thorough radiographic examination is the only reliable method of confirming the clinical diagnosis. The congenital fractures are either

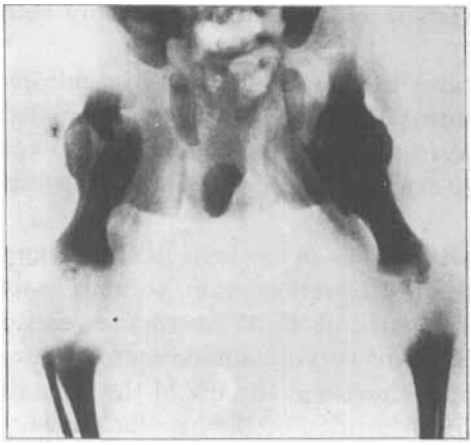


Fig. 11.
(Case 17, boy K. W.) Multiple malformations. Recent fractures of both femora (intra-uterine), with exuberant callus immediately after birth. Postero-medial angulation at the upper third and congenital dislocation of both hips. The boy also had spina bifida with meningo-myelocele and hydrocephalus.

solid with marked angulation or show some callus immediately after birth or a definite pseudarthrosis. *Camurati* describes three different radiographic types. In *type I* there is bone apposition and thickening at the maximum of the angulation; it may remain solid forever when properly treated. Probably *Kitchin*'s "atypical case of infantile cortical hyperostosis" (1951) belongs to this group. *Type II* shows thickening of the inside of the cortex and narrowing of the medullar canal but no apposition; it is less solid and tends to early fracture and pseudarthrosis. In *type III* there is either found some thickening of the inner cortex of the upper fragment only, whereas the lower fragment has very thin walls; or both fragments are very thin-walled with pointed sclerotic ends placed at a rather acute angle in a typical pseudarthrosis. The latter has a remarkable bad prognosis.

A case of osteogenesis imperfecta congenita was even diagnosed radiographically in utero five weeks ante partum. In spite of a proper size of the uterus in keeping with the ninth month of pregnancy and a distinct heart sound there were only found very faint shadows of the bones of the foetus, and malformation was suspected (*Hortemo,* 1953).

Traumatic fractures are easily ruled out when x-rayed immediately after birth; there is found a fresh fracture with hematoma but no callus. After six days callus is abundant, and after two weeks union is solid.

Inflammatory, scorbutic and rachitic lesions show typical radio-

graphic characteristics, and the changes rapidly subside after proper treatment.

Multiple fractures in infants, fractures in children with blue sclerotics or skin pigmentations, and fractures associated with malformations should always be suspected of being due to congenital bone deficiency, and x-ray examination of the whole skeleton should be undertaken.

Laboratory methods are of no help in the diagnosis of congenital fractures. The values of serum calcium, serum phosphorus and serum phosphatase are normal, and the Wassermann reaction is negative.

Histologically is found varying amounts of fibrous connective tissue between the osseous lamellas at the site of the maximal bending, sometimes with lacunar absorption of the osseous tissue.

TREATMENTS AND PROGNOSIS

The prognosis of these lesions depends on the type of fracture or angulation, the age of the patient when treatment begins, and which method of treatment is chosen. The results can only be evaluated after a very long observation period.

The lower leg.

As previously mentioned *posterior angulations* have a fair prognosis, possibly because the "bow-string activity" (*Birkett*) of the extensor muscles is not so pronounced as that of the hamstring muscles. *Heymann & Herndon* and *Miller* advise against osteotomy or grafting in these cases and advocate conservative treatment consisting in massage and stretching of the soft tissues which hold the foot in calcaneous position. Later a toe-to-groin brace is used with a doubled-back leather cuff capable of being tightened posteriorly at the site of the angulation with straps and buckles; there should be a stop joint at the ankle and rigid lateral uprights. This brace should be applied when the child is 5 months of age, before it begins weight-bearing, and it should be continued until the angulation is corrected by growth and the calcaneous deformity is eliminated. 1–2 cm shortening must be expected.

In *Dawson's* case a corrective osteotomy was performed and the fragments were fixed by a plate with four screws. Soon after a fracture occurred at the upper screw, but the eventual result became satisfactory.

The *anterior angulations,* when possible, should be treated at first conservatively for several years, at least for 5–8 years (*Camurati; Scott*; *Boyd*; *Moore*). A plaster cast or a brace should be applied which

should be changed as the child grows up. Radiographs should be taken every two months, and if necessary manipulative reduction should be carried out.

These conservative measures should also be tried in the treatment of fractures and pseudarthrosis. The older methods of injecting iodine or whole blood at the site of fracture have proved of no value. Ignipuncture and radiation are ineffective. Continuous traction should not be applied in cases of fracture and pseudarthrosis, but may improve congenital bending of the lower legs (*Freund,* 1936).

Of the operative methods bone suture and bands as well as plates and screws have invariably proved disappointing. Osteotomy with correction of the alignment may be successful but not until the child is at least 6 years of age. Resection most often will be insufficient as a too large area of bone is involved so that the eventual shortening becomes too conspicuous. It was recommended by *Hayashi & Matsuoka* (1922). Occasionally, excochleation of a localized cyst may be sufficient, but *Compére* (1936) found refractures even after post-operative immobilization for 12 months.

Bone grafting is the method of choice. A complete substitution of the defective bone tissue by healthy bone must be aimed at (*Aegerter,* 1950). Repeated graftings may be necessary.

It appears that some of the various techniques successfully used for bone transplantation in ordinary fractures are not suitable in the treatment of congenital fractures and pseudarthrosis.

Camurati (1930) used autotransplants from the patient's healthy tibia, either free massive (Albee) or laminar, one-piece (Froelich) and multiple (Codivilla) osteoperiosteal grafts, or peduncular transplants, permanent (Wolf-Müller) or temporary (Reichel). He had only 5 satisfactory results in 27 cases.

Henderson (1925) using similar methods had malunion in everyone of 7 operated children under 5 years of age. *Hallock* (1938) suggested an improved technique on the basis of the accepted theories of bone regeneration. After having removed all the pseudarthrotic tissue he chiselled out a cleft in the bone down to the medullar cavity in the bone ends well proximally and distally from the site of the pseudarthrosis. It was filled with small bone chips from the iliac crest, and the periosteum and the soft tissue layers were closed. Afterwards a plaster cast was applied for 10–12 weeks and then a walking cast until union was radiographically safe.

McFarland (1940) took a long solid graft from the healthy tibia and inserted it as a chord on the back of the pseudarthrotic tibia at a

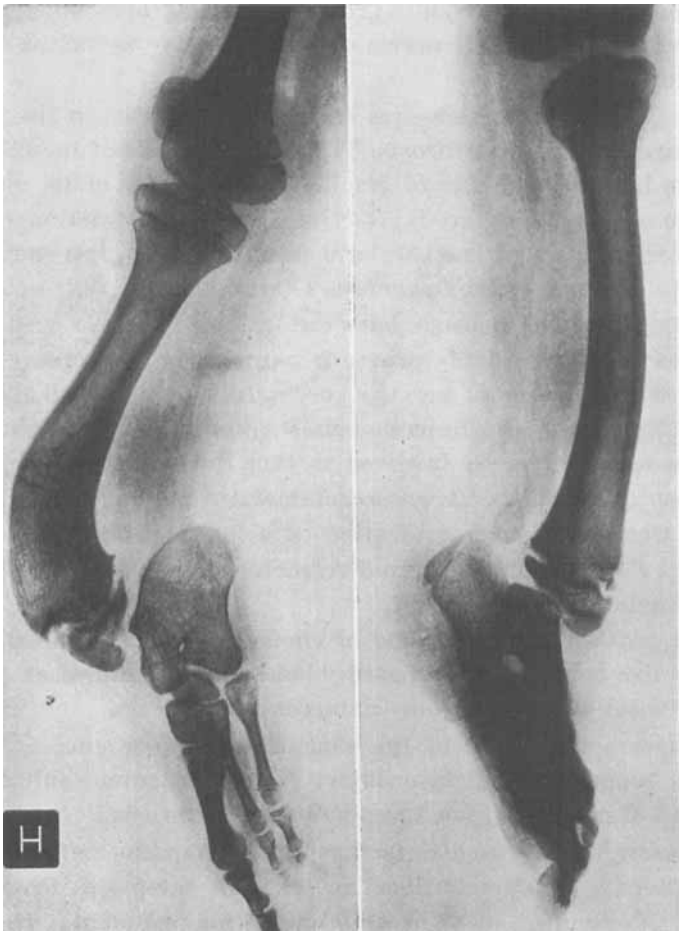


Fig. 12.
(Case 18, girl H. D.) Multiple malformations. Seen at 8½ years of age. Antero-medial angulation of both tibiæ, absence of both fibulæ. Absence of two right toes and one left toe. Dislocation of the left knee. Shortening of the right ulna and antero-lateral angulation of the right radius with dislocation in the elbow. Absence of three right fingers. Both lower legs had to be amputated.

proper distance upwards and downwards from the angulation, "by-pass" technique. He reported solid union in 7 out of 9 cases after follow-up examination (1951), but shortening usually was marked.

Watson-Jones (1946) thinks that angulation of the tibia should not be maintained, as a second pseudarthrosis is liable to develop at the site of impaction of the lower end of the "by-pass" graft if weight bearing passes in front of the ankle joint. He favours the method of *Boyd* (1941) who suggested long dual onlay grafts from the mother

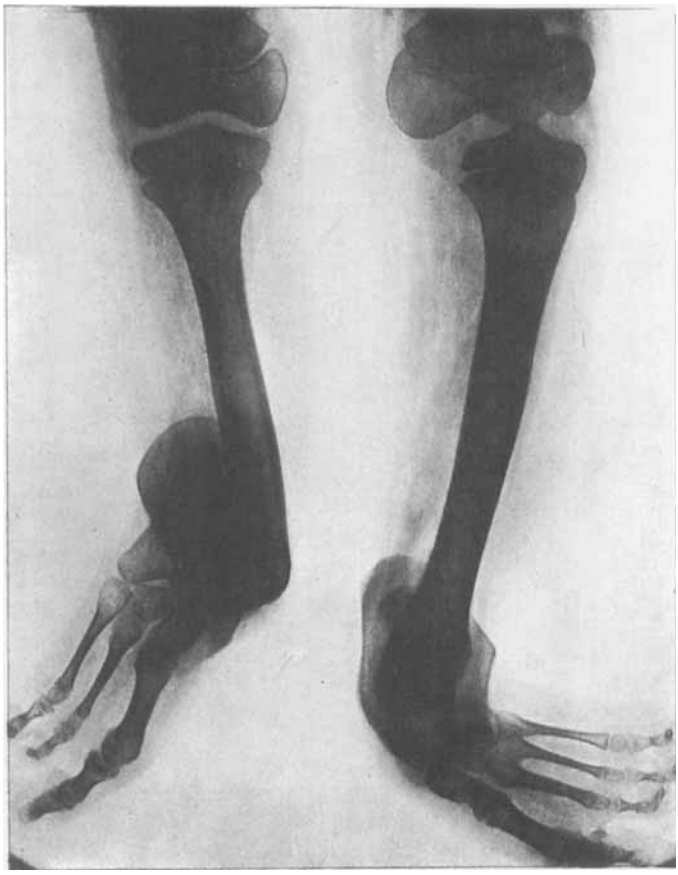


Fig. 12.

or the healthy tibia of the child, fixed by two vitallium screws at both fragments of the pseudarthrosis, with bone chips between the ends. Afterwards a brace should be carried for several years until a normal medullary canal had been reconstructed. The final results were, however, not quite satisfactory as 4 out of 7 cases had refractures (*Boyd & Fox*, 1948).

Wilson (1941) performed a "two-stage" transplantation of the fibula of the leg with pseudarthrosis of the tibia where the latter only was affected. The fibula was severed, first at the upper end and impacted, and about 4 weeks later the procedure was repeated at the lower end of the tibia. Plaster cast was applied for 12–13 weeks and afterwards a walking cast for 3 months. He was successful in 4 out of 5 cases with congenital pseudarthrosis and believed that healing was made possible because of the absolute immobilisation obtained

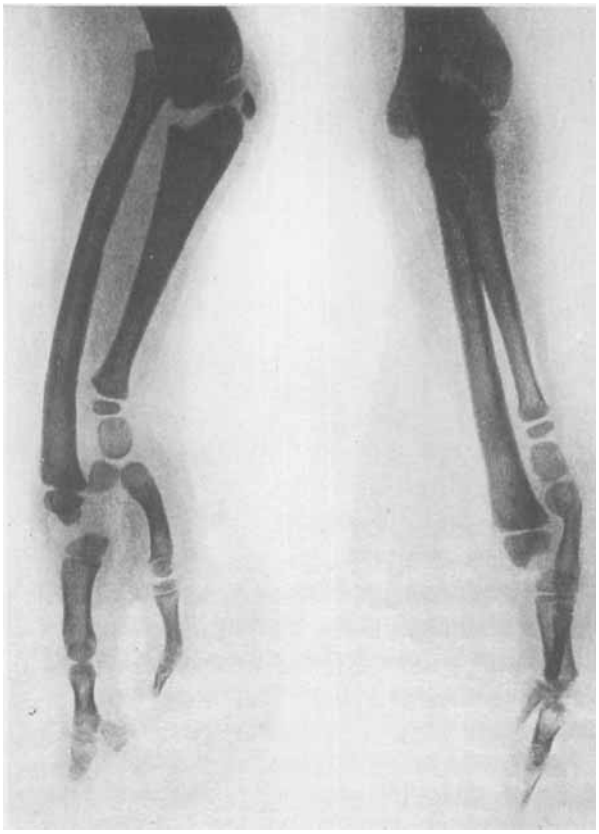


Fig. 12.

through the splint effect of the fibula. *Moore* (1949) preferred delayed autogenous bone grafts from the healthy tibia, the fibula or the iliac crest. The grafts were elevated and freed of attachments and afterwards replaced in their natural bed for about 3 weeks. Thereafter, they were removed and implanted in the fragments of the pseudarthrosis after complete excision of the fibrous tissue and the sclerotic bone ends. The fragments were transfixed with two Kirschner wires passing through each of the two ends, avoiding the epiphyseal lines. They were attached to an *Abbott*'s bone lengthening apparatus. The wires and a plaster cast from toes to groin were maintained for about 4–6 months until a medullary cavity appeared on the radiograph. Then a double-bar caliper brace from ankle to groin with a pelvic band and a full length tibial leather cuff was used for two years.

X-ray control was made every two months. 5 cases of congenital pseudarthrosis were successfully treated but observation time is as yet

too short. *Aegerter* (1950) recommends this method of bringing in a new set of osteoblasts from an uninvolved area.

The femur.

Very few authors mention the treatment of congenital bowing and shortening of the femur. Because of the risk of dislocation of the hip the child should not be allowed to stand up or walk early. A plaster spica to the waist may be necessary or a walking splint with support on the tuberosity of the ischium. Osteotomy and bone lengthening should be undertaken as late as possible, as there is a tendency to spontaneous improvement.

Ollerenshaw (1925) reported a remarkable operation on a girl of 17 years of age with angulation of her left femur and 4½ inches' shortening. The short femur was freed of attachments and sawed through; then traction was applied for some minutes until a gap of 1½ inches was formed. Now the normal femur was incised; 1½ inches of the bone was sawed out from the middle of the diaphysis and the ends were fixed together by a Lanes plate and screws. The bone cylinder from the right femur was then inserted in the gap in the short femur and fixed by another Lanes plate. Plaster cast from toes to the curvatures was applied for some weeks, and the eventual result was very satisfactory.

The forearm.

Lee (1933–34) advocates conservative measures until the child is near puberty. Then partial resection may be considered, whereas bone grafting is not recommended and possibly not necessary.

Even if the prognosis of congenital fractures must still be considered very bad, or at least very uncertain, solid union *can* take place eventually, and great patience must necessarily be displayed by the patient as well as by the orthopoedic surgeon. With improved technique and better understanding of the true nature of these lesions amputation should be more and more seldom resorted to.

In some cases, however, with severe congenital defects it is just as well to amputate early with conservation of everything of value. Children will quickly get accustomed to a prosthesis and learn to compensate for their congenital defect.

TABLE I

Case no. record	Sex	Lesion	Age when first noticed	Age when admitted in hospital	Angulation (Other anomalies)	Shortening (dimple)	Pseudarthrosis	Blue scleræ	Pigmentation or nævi
1 OH 631/38	F	Bending of left lower leg at 3 months, fracture of left tibia at 14 months, subsequent pseudarthrosis	3 mths.	5 mths.	Anterolateral (pes varus adductus)	2 cm (—)	+	—	—
2 OH 786/30	F	Bending of right lower leg at birth, fracture at 2 years, subsequent pseudarthrosis	new-born	2 years	Anterior (—)	Slight (—)	+	—	—
3 OH 670/50	M	Fracture at lower third of right lower leg at 2 years, subsequent pseudarthrosis	2 years	58 years	Anterior (talipes equinus kypho-scoliosis)	20 cm (—)	+	—	+
4 OH 1779/40	M	Bending of right lower leg at 14 months, fracture of right lower leg at 5 years, subsequent pseudarthrosis	14 mths.	5 years	Anterior (—)	5 cm (—)	+	—	—

TABLE I

First radiograph	Treatment	Follow-up after	Latest radiograph	Result (Clinical diagnosis)
Oblique fracture of the middle of left tibia, 140° antero-lateral angulation	a) Reduction and plaster cast for 5 months b) Brace for 5 years c) Posterior cortical Albee transplant at 5 years d) Osteotomy and two full grafts from mother, vitallium screws and bone chips at 11 years e) Thomas' splint, caliper brace	15 years	Solid healing, 20° anterior angulation, talipes calcaneus	4 cm shortening, solid healing, function normal. Satisfactory result (Congenital pseudarthrosis of tibia)
Fracture of lower third of tibia and fibula. Sclerotic, pointed ends, displacement, 150° anterior angulation	a) Caliper brace with boot	8 years	Not available	Walks with a caliper brace. Eventual result unknown (Congenital pseudarthrosis of lower leg)
Not available	a) Caliper brace	56 years	Not available	Walks with a caliper. Receives disablement pay (Neurofibromatosis)
140° anterior angulation at lower third of right lower leg (congenital fracture sequelæ?)	a) Caliper brace and boot for 6 years, thereafter b) pin traction for 3 weeks c) followed by osteosynthesis: vitallium plate, cortical graft and bone chips d) Full graft on-lay after 4 months e) Caliper brace for 4 years f) Amputation of right lower leg at 11 years	17 years	Pseudarthrosis of right tibia and fibula at lower third. Fracture of graft. Marked displacement, bone absorption and shortening	Walks well with prosthesis after amputation (Congenital pseudarthrosis of lower leg)

TABLE I (cont.)

Case no. record	Sex	Lesion	Age when first noticed	Age when admitted in hospital	Angulation (Other anomalies)	Shortening (dimple)	Psendarthrosis	Blue scleræ	Pigmentation or nævi
5 OH 7389/51	F	Fracture of right lower leg at 18 months subsequent pseudarthrosis	18 mths.	18 mths.	Postero-medial (—)	$4^1/_2$ cm (—)	+	—	—
6 OH 422/26	F	Pseudarthrosis of left lower leg at birth	new-born	2 years	Anterior (talipes equinus)	4 cm (—)	+	—	+
7 OH 6292/39	F	Fracture of left lower leg at birth	new-born	3 mths.	Postero-medial (—)	$1^1/_2$ cm (—)	+	—	—
8 OH 3928/43	M	Fracture of left lower leg at 10 months	10 mths.	13 mths.	Anterolateral (—)	4 cm (—)	+	—	—

TABLE I (cont.)

First radiograph	Treatment	Follow-up after	Latest radiograph	Result (Clinical diagnosis)
Fracture at lower third of right tibia and fibula, posterior angulation	a) Repeated reductions and plaster cast for 2 weeks b) Osteosynthesis with vitallium plate after 4 weeks c) Cortical graft from left tibia after 6 months, leather brace afterwards d) Osteosynthesis with vitallium plate after 2½ years	4½ years	Pseudarthrosis of right tibia and fibula at lower third. Pointed, sclerotic ends	Eventual result unknown (Congenital pseudarthrosis of lower leg)
Not available	a) Plaster cast for 13 months b) Osteotomy and wire suture c) Caliper brace; the patient did not want operation	37 years	Pseudarthrosis of left tibia and fibula	Walks with a caliper, 11 cm shortening, scar and sinus after osteotomy Receives disablement pay (Neurofibromatosis)
Fracture of lower third of left tibia and fibula, slight postero-medial angulation	a) Reduction at 2 months b) Repeated reductions three times, plaster casts	13 years	Solid healing and perfect alignment	Satisfactory result, normal function, 1½ cm shortening, slight scoliosis (Intra-uterine fracture of lower leg)
Anterior angulation of left tibia and fibula with bone cysts at lower third	a) Excochleation of fibrous tissue from the cysts at 13 months b) Graft of os purum at 2 years c) Osteotomy at 6 years d) Bone pegging with a piece of fibula at 7½ years e) Beck's drillings at 9 years, and caliper brace f) Osteosynthesis with Küntscher nail planned	17 years	Marked anterior angulation, fibula not healed, a few cysts	Satisfactory result, solid healing, 5 cm shortening. Walks with a caliper (Ostitis fibrosa cystica localisata)

TABLE I (cont.)

Case no. record	Sex	Lesion	Age when first noticed	Age when admitted in hospital	Angulation (other anomalies)	Shortening (dimple)	Pseudarthrosis	Blue scleræ	Pigmentation or nævi
9 OH 1985/51	F	Bending of left femur at birth	newborn	newborn	Anteralateral (dislocation of left hip)	4 cm (— at the middle of femur laterally)	—	—	—
10 OH 2668/38	F	Bending of both femora at birth	newborn	2 weeks	Lateral (contracture at the knees)	3 cm (—)	—	—	—
11 A 563/43	M	Bending of both femora at birth, fracture of right malleolus at 1 year, fracture of right lower leg at 18 months	newborn	newborn	Left femur: lateral at upper third right femur: medial, at lower third	Both sides 2 cm (—)	—	—	—
12 DLB 753/43	F	Bending of both femora and both lower legs at birth	newborn	1 mths.	Both femora: anterior, both lower legs: anterior	Both sides 2 cm (—)	—	—?	—
13 OH 1029/50	M	Bending of right forearm (at birth?)	2 years	6 years	Anterolateral (kyphosceoliosis, spina bifida)	1 cm (—)	—	—	—
14 A 811/45	F	Fractures of both femora and right clavicle at birth	newborn	newborn	Both femora: lateral (spina bifida, meningocele, hydrocephalus, talipes equinus)	Both sides 3 cm (—)	Both sides fracture	—	—

TABLE I (cont.)

First radiograph	Treatment	Follow-up after	Latest radiograph	Result (Clinical diagnosis)
150° antero-lateral angulation at upper third of left femur, no callus	a) No weight bearing for 22 months b) Walking splint c) Corrective osteotomy planned when patient is 4 years	3 years	80° coxa vara, marked lateral angulation at upper third of left femur, epiphyseal line oblique	9 cm shortening, femur solid, walks with splint, observation time too short (Intra-uterine fracture of femur)
150° lateral angulation of both femora, posterior thickening of the cortex, no callus	a) No weight bearing, redressive manipulations of the knees	5 years	Slight bowing of both femora, normal bone structure	Child walks better, only 10° contracture of the knees, feet can be placed with parallel axes. Result satisfactory (Intra-uterine fractures of femora)
150° lateral angulation of left femur, 160° medial angulation of right femur. Cortical thickening at concavity	No treatment	3 years	Slight lateral bowing of left femur, normal bone structure	Bilateral talipes plano-valgus, bones solid at present, but observation time too short (Osteogenesis imperfecta)
Faint bone shadows, thin compacta, fracture sequelæ at lower third of both femora, no callus	a) No weight bearing	—	Not available	Observation time too short (Osteogenesis imperfecta)?
Distal half of right ulna deficient, lateral angulation of radius	a) Splint operation suggested but postponed as child is psychically abnormal	8½ years	Increasing angulation of radius with dislocation of capitulum. Cysts and dysplastic bone in ulna	Function not impaired, movements in elbow°: 180°/65°, shortening 2½ cm (Neurofibromatosis)
Fractures of both femora in upper third, no callus	No treatment	—	Not available	Result unknown (Multiple congenital malformations)

TABLE I (cont.)

Case no. record	Sex	Lesion	Age when first noticed	Age when admitted in hospital	Angulation (other anomalies)	Shortening (dimple)	Pseudarthrosis	Blue scleræ	Pigmentation or nævi
15 B 1227/37	F	Bending of both femora and left lower leg, fresh fracture of right lower leg and clavicle at birth	newborn	newborn	Both femora: lateral both lower legs: anterior (—)	Both sides 3 cm (—)	Both femora solid, fracture of right lower leg	+	—
16 OH 12324/51	M	Bending of left femur and both lower legs at birth, subsequent pseudarthrosis of right tibia	newborn	2 days	Left femur: lateral, both lower legs: anterior (absence of right fibula, right and left toes, right talipes equinus)	Right lower leg: 5 cm (+ on both lower legs anteriorly)	Right tibia — other bones: solid	—	—
17 OH 3411/47	M	Fractures of both femora at birth	newborn	1 mth.	Both femora: posteromedial (spina bifida, dislocation of both hips, hydrocephalus)	Both sides 4 cm (—)	Healing fractures	—	—
18 OH 7044/44	F	Bending and other deformities of both lower legs and right forearm	newborn	16 mths.	Both tibiæ: anterior, right radius: lateral (absence of both fibulæ, several toes and fingers, talipes equinus, disloc. of knees)	Both sides 5 cm	—	—	—

TABLE I (cont.)

First radiograph	Treatment	Follow-up after	Latest radiograph	Result (Clinical diagnosis)
Faint bone shadows, compacta thin, angulation of both femora and left lower leg, fracture of right lower leg, no callus	No treatment	—	Not available	Result unknown (Osteogenesis imperfecta)
Medial angulation of left femur, anterior angulation of left lower leg, marked anterior angulation of right tibia, right fibula absent	a) Manipulation of the feet b) Cuneiform osteotomy of right tibia at 2 months, tenotomy, plaster cast c) Repeated reduction, caliper brace	2 years	Fracture at the site of the osteotomy, pseudarthrosis with 90° angulation	Observation time too short (Multiple congenital malformations)
Oblique fractures of both femora at upper third, postero-medial angulation, marked callus	No treatment	1/2 year	Not available	Child died at 7½ months (Multiple congenital malformations)
Marked anterior angulation of both tibiæ, absence of both fibulæ and several toes and fingers. Dislocation of radius at the elbow	a) Plaster casts b) Leather brace for 7 years c) Amputation of both lower legs at 9 years (above the knee)	8 years	Increasing deformity of both lower legs. Rudiment of lower end of left fibula, dislocation of left knee	Child walks well with two prostheses (Multiple congenital malformations)

ANALYSIS AND FOLLOW-UP EXAMINATION OF 18 CASES OF CONGENITAL ANGULATIONS AND FRACTURES OF THE EXTREMITIES

It was possible to find 18 cases among the patients in the Orthopædic Hospital of Copenhagen and in the Obstetric Departments of the University Hospital of Copenhagen, 14 patients could be followed-up from 2 to 58 years after the first occurrence of the lesion, but only in a few cases the final result could be evaluated.

There were 7 males and 11 females. In 12 cases the lesion was noticed already at birth, in 2 cases during the first year, in 3 cases during the second year, and in 1 case during the third year.

All the lesions are considered due to deficient bone development during early foetal life, in conformity with the theories and experimental findings of *Duraiswami.*

Instead of detailed case reports a brief synopsis of all the cases and their treatment is given in Table I.

Morphologically their characteristics are shortening and angulation of the diaphysis with or without subsequent fracture and pseudarthrosis. They are grouped as follows:

(1) lesions of the tibia	1 case	(No. 1, Fig. 1)
(2) lesions of the tibia and the fibula	7 cases	(No. 2–3–4–5–6–7–8, Fig. 2–3–4–5)
(3) lesions of the femur or both femora	4 cases	(No. 9–10–11–12–, Fig. 6–7–8)
(4) lesions of the forearm	1 case	(No. 13, Fig. 9)
(5) lesions of long bones and other bones	2 cases	(No. 14–15)
(6) multiple malformations with or without absence of bones	3 cases	(No. 16–17–18, Fig. 10–11–12)

The clinical diagnoses were:

(a) osteogenesis imperfecta	3 cases	(No. 11–12–15, Fig. 8)
(b) neurofibromatosis	3 cases	(No. 3–6–13, Fig. 9)
(c) ostitis fibrosa cystica localisata	1 case	(No. 8, Fig. 5)
(d) congenital pseudarthrosis without blue scleræ or pigmentations	4 cases	(No. 1–2–4–5, Fig. 1–2–3)
(e) intra-uterine fracture sequelæ	3 cases	(No. 7–9–10, Fig. 4–6–7)
(f) multiple malformations	4 cases	(No. 14–16–17–18, Fig. 10–11–12)

Lesions of the lower leg.

The lower leg was involved in 12 cases, 4 on the right leg, 4 on the left leg and 4 bilateral. As a rule both the tibia and the fibula were affected. In 10 out of 11 cases the site of the lesion was at the lower third.

Anterior angulation was found in 10 cases (4 bilateral). 3 remained solid, 2 had subsequent fractures which eventually healed, and 5 had pseudarthrosis.

Posterior angulation was seen in 2 cases. One fractured but healed afterwards; the other had still pseudarthrosis after 4½ years.

It has been claimed that posterior angulation has a better prognosis than anterior angulation. Pseudarthrosis may, however, occur also in posterior angulation as seen in case No. 5. On the other hand anterior angulation may occasionally remain solid (case No. 18). The tendency towards malunion, therefore, cannot solely be attributed to the action of the strong hamstring muscles.

Amputation had to be performed in 2 cases. One of them (case No. 18) had multiple malformations and had no chance of recovery although conservative measures were tried for 8½ years. The other patient (case No. 4) had two unsuccessful operations made after 6 years of conservative treatment, and shortening had already become very marked.

In the other cases no general principles have been followed, and no special operative method can be recommended on the basis of such few cases. It seems, however, advisable to be conservative for at least 7–8 years, as suggested by other authors. Regular and frequent roentgen examinations, manipulative reduction of the fracture, repeatedly when necessary, and immobilization in plaster casts and caliper braces is advocated. In one case (No. 7) a satisfactory result was obtained without operation at all, and in another case (No. 1) operation was unsuccessful after 5 years but successful after 11 years of age.

As a whole, prognosis seems to be very discouraging in congenital lesions of the lower leg. Solid union may be obtained, but usually a marked shortening must be anticipated. It is, however, possible that the results will improve when newer operative methods are tried, aiming at substitution of all defective bone tissue by healthy bone, for instance by *Hallock*'s or *Moore*'s technique.

Lesions of the femur.

There were 8 cases of angulation of the femur. Only one of them (case No. 17) had recent fractures with callus. Delivery had been spontaneous and trauma could be ruled out. The child had congenital dislocation of the hips and spina bifida with meningocele so that deficient bone development with multiple malformations was most likely the causative factor. In all the other cases there was found a solid angulation of the diaphysis, but one cannot be sure that this condition will be lasting.

As a rule bilateral angulation was found, and only in two cases was the lesion unilateral. In one of these, shortening was as much as 9 cm at the age of 3 years.

The site of the angulation was either at the upper or the lower third of the diaphysis. In the bilateral lesions the angulations were not necessarily symmetrical.

Treatment was conservative for the first years. In the unilateral lesions there is a great risk of scoliosis and coxa vara. Corrective osteotomy and bone lengthening operations must therefore be considered later. In the meantime a walking splint should be applied as soon as the child commences to stand upright.

One of the bilateral lesions (case No. 10) improved spontaneously and disablement was only slight after 5 years. In the rest of the cases observation time has been too short to predict anything about the prognosis.

Lesions of the forearm.

Only 2 cases were found. The right forearm was involved in both cases. There was considerable shortening of the ulna and increasing angulation of the radius with dislocation of the capitulum radii behind the lower end of the humerus. The function of the elbow joint did not suffer much. One case (No. 18) had severe malformation of the hand and fingers.

Corrective osteotomy may be necessary later, whereas splints are not likely to be of any use. The angulations seem to remain solid, and movements in the elbow should be encouraged.

The final result cannot yet be evaluated.

SUMMARY

The various etiologic factors attached to congenital angulations and fractures are discussed, and it is concluded that these lesions are due to defective ossification involving a large area of the diaphysis.

Angulation, fracture and pseudarthrosis are considered different forms of the same bone lesion. The characteristic symptoms and signs are described, and it is emphasized that radiographic examination is necessary for the diagnosis.

Several methods of treatment are mentioned. As a general principle, only manipulative reduction and immobilization should be preferred during the first 7 years. Operation should aim at a complete substitution of the defective bone tissue by autografts of healthy bone. Subsequent immobilization in plaster casts and caliper braces must be

continued, under frequent roentgen control, for many months until solid union is established in lesions of the lower leg. Corrective osteotomy may be necessary in angulations of the femur and the forearm.

18 cases are reported.

RESUME

Il est discuté des différents facteurs étiologiques qui se rattachent aux angulations et aux fractures congénitales et il en est conclu que ces lésions sont dues à une ossification défectueuse s'étendant sur une grande surface du diaphyse.

L'angulation, la fracture et la pseudarthrose sont considérées comme des formes différentes de la même lésion osseuse. Les symptômes caractéristiques et les signes de la maladie sont décrits et il est souligné que l'examen radiologique est nécessaire pour poser le diagnostique.

Plusieurs méthodes de traitement sont mentionnées. D'une manière générale, il convient de donner la préférence à un traitement qui comporte uniquement la réduction manipulative et l'immobilisation pendant les premiers 7 ans. L'opération doit viser à remplacer le tissu osseux défectueux par des autogreffes d'os sain. Une immobilisation subséquente dans des moules de plâtre et des bandages caliper doit être maintenue, sous contrôle radiologique fréquent, pendant de longs mois jusqu'à ce qu'une consolidation solide soit établie, lorsqu'il s'agit de lésions dans la partie inférieure de la jambe. Une ostéotomie corrective peut être nécessaire dans les angulations du fémur et de l'avant-bras.

18 cas sont rapportés.

ZUSAMMENFASSUNG

Die verschiedenen ursächlichen Faktoren angeborener Knickungen und Brüche der Knochen werden besprochen. Man gelangt zu dem Schluss, dass diese Schädigungen ihre Ursache in einer weitgehenden, mangelhaften Verknöcherung der Diaphyse haben.

Knickung, Bruch und Pseudarthrose werden als verschiedene Formen der selben Knochenschädigung angesehen. Die charakteristischen Symptome werden beschrieben und es wird hervorgehoben, dass die Röntgenuntersuchung zur Stellung der Diagnose notwendig ist.

Einige Behandlungsmethoden werden erwähnt. Grundsätzlich sollte während der ersten 7 Jahre gedeckte Reposition und Fixation als Behandlung vorgezogen werden. Die Operation sollte eine vollständige Erstattung des fehlerhaften Knochengewebes mittels Verpflanzung von eigenem, gesunden Knochen anstreben. Die folgende Ruhig-

stellung im Gipsverband und mittels Apparaten muss unter ständiger Röntgenkontroll viele Monate hindurch aufrecht erhalten werden bis eine solide Vereinigung in Fällen von Schädigung des Unterschenkels stattgefunden hat. Korrigierende Osteotomien können sich bei Knickungen des Femurs und Unterarms als notwendig erweisen.

18 Zufälle sind beschrieben.

REFERENCES

Abesser, E.-W.: Intrauterin erworbene Osteomyelitis? Fortschr. Roentgenstr. 78: 213, 1953.

Aegerter, E. E.: The possible relationship of neurofibromatosis, congenital pseudarthrosis and fibrous dysplasia. J. Bone & Joint Surg. 32 A: 618–626, 1950.

Bentzon, P. K.: Roentgenological and experimental studies on the pathogenesis of the dyschondroplasia (Ollier's disease). Acta Radiologica 3: 89, 1924.

Birkett, A. N.: Note on pseudarthrosis of the tibia in childhood. J. Bone & Joint Surg. 33 B: 47–49, 1951.

Bludau, F. M.: Beitrag zum Capitel der intrauterin erfolgten Fracturen bei Neugeborenen. Inaug. diss. Berlin 1893.

Boyd, H. B.: Congenital pseudarthrosis. Treatment by dual bone grafts. J. Bone & Joint Surg. 23: 497–515, 1941.

Boyd, H. B. & K. W. Fox: Congenital pseudarthrosis. Follow-up study after massive bone-grafting. J. Bone & Joint Surg. 30 A: 274–283, 1948.

Browne, D.: Congenital deformities of mechanical origin. Proc. Royal Soc. Med. 29 II: 1409–1431, 1936.

Büungner, O. v.: Ueber intra partum entstandene Unterschenkelfrakturen. Arch. f. klin. Chir. 41: 174–186, 1891.

Camurati, M.: Le pseudarthrosi congenite della tibia. Chir. d. Org. di Movimento 15: 1–162, 1930.

Clark, D. H., Lumberton, N. C. & W. R. Mathews: Simultaneous occurrence of von Recklinghausen's neurofibromatosis and osteitis fibrosa cystica. Surgery 33: 434–443, 1953.

Compére, E. L.: Localized osteitis fibrosa in the new-born and congenital pseudarthrosis. J. Bone & Joint Surg. 18: 513–523, 1936.

Dawson, G. R. jr.: Intra-uterine fractures of the tibia and fibula. J. Bone & Joint Surg. 31 A: 406–408, 1949.

Ducroquet, R.: A propos des pseudarthroses et inflexions congénitales du tibia. Mém. de l'Acad. Chir. 63: 863–868, 1937.

Duraiswami, P. K.: Experimental causation of congenital skeletal defects and its significance in orthopedic surgery. J. Bone & Joint Surg. 34 B: 646–698, 1952.

Eliason, E. L.: Pathological fractures. Surg., Gunec. & Obst. 56: 504–521, 1933.

Evans, W. A.: Recurrent fracture. Surg., Gynec. & Obst. 74: 204–219, 1942.

Fraser, J.: Fragilitas ossium tarda. Brit. J. Surg. 22: 231–240, 1934–35.

Freudenberg, E., Roulet, F. & R. Nicole: Kongenitale Infektion mit Coxsackie-Virus. Ann. Pædiatr. 178: 150–161, 1952.

Freund, E.: Congenital defects of femur, fibula and tibia. Arch. Surg. 33: 349–391, 1936.

Frølich, T.: Lues congenital præcox. Nordisk Lærebog i Pædiatri II: 226–233, 1941.

Granzow, J.: Intrauterin erworbene, komplizierte Spontanfraktur des Unterschenkels durch amniotische Stränge. Z.bl. f. Gynäk. 54: 2699–2702, 1930.

Green, ? : Frische Oberschenkelfraktur bei Neugeborenen. Z.schr. f. Geburtsh. u. Gynäk. 41: 522–524, 1899.

Gurlt, E.: Über intrauterine Verletzungen des fötalen Knochengerüstes vor und während der Geburt. Monatsschr. f. Geburtskunde 9: 321–349; 401–436, 1857.

Hallock, H.: The use of multiple small bone transplants in the treatment of pseudarthrosis of the tibia. J. Bone & Joint Surg. 20: 648–660, 1938.

Harnett, W. L.: Two cases of osteogenesis imperfecta with blue sclerotics in natives of India. Brit. J. Surg. 22: 269–273, 1934–35.

Hauser, F.: Über angeborene Rachitis. Ann. Pædiatr. 180: 299–306, 1953.

Hayaschi, K. & M. Matsuoka: Ueber intra partum entstandene Unterschenkelfrakturen. Arch. f. klin. Chir. 98: 417–427, 1912.

Henderson, M. S.: Pseudarthrosis of the tibia in children. J. Bone & Joint Surg. 7: 340–354, 1925.

Henning, M.: Ein Fall von kongenitaler doppelseitiger Oberschenkelfraktur mit Nabelschnurumschlingung bei einem ohne Kunsthilfe geborenen Kinde. Inaug. diss. Berlin 1900.

Heymann, C. H. & C. H. Herndon: Congenital posterior angulation of the tibia. J. Bone & Joint Surg. 31 A: 571–580, 1949.

Hortemo, T.: Osteogenesis imperfecta congenita diagnostisert intrauterint. Nord. Med. 49: 574–575, 1953.

Hunter, D. & Ph. Wiles: Dyschondroplasia (Ollier's disease) with report of a case. Brit. J. Surg. 22: 507–519, 1934–35.

Hölder, H.: Intrauterine Femurfraktur bei Spontangeburt in Scheidellage. Zbl. f. Gynäk. 41: 127–130, 1917.

Kitchin, I. D.: An atypical case of infantile cortical hyperostosis. J. Bone & Joint Surg. 33 B: 248–250, 1951.

Kramer, O.: Ein Fall angeborener (intrauteriner) complicirter Fractur des Unterschenkels. Münch. med. Wschr. 1238–1239, 1900.

Kugelmann, J.: Über symmetrische Spontanfrakturen unbekannter Genese beim Säugling. Ann. Pædiatr. 178: 177–189, 1952.

Lee, M.: Congenital absence of the fibula with associated deformity of the femur. Proc. Royal Soc. Med. 27: 1257, 1933–34, II.

– Congenital absence of ulna with associated deformity of radius. Proc. Royal Soc. Med. 27: 1268–1269, 1933–34, II.

Lewin, Ph.: Arthrogryposis multiplex congenita. J. Bone & Joint Surg. 23: 630–636, 1925.

Madsen, E. T.: Fractures of the extremities in the newborn. Acta Obst. & Gynec. Scand. 34: 41–74, 1955.

Madsen, V.: personal communication. 1953.

McCarroll, H. R.: Clinical manifestations of congenital neurofibromatosis. J. Bone & Joint Surg. 32 A: 601–617, 1950.

McFarland, B.: "Birth fracture" of the tibia. Brit. J. Surg. 27: 706–712, 1940.

– Pseudarthrosis of the tibia in childhood. J. Bone & Joint Surg. 33 B: 36–46, 1951.

Middleton, D. S.: The Pathology of congenital genu recurvatum. Brit. J. Surg. 22: 696–702, 1934–35.

– Studies on prenatal lesions of striated muscle as a cause of congenital deformity. Edinburgh Med. J. 41: 401–442, 1934.

Miller, B. F.: Congenital posterior bowing of the tibia with talipes calcaneo-valgus. J. Bone & Joint Surg. 33 B: 50–55, 1951.

Miller, G.: Die Knochenveränderungen bei der Neurofibromatosis Recklinghausen, Fortschr. Roentgenstr. 78: 669–689, 1953.

Moore, B. H.: Some orthopædic relationships of neurofibromatosis. J. Bone & Joint Surg. 23: 109–140, 1941.

Moore, J. R.: Delayed autogenous bone graft in the treatment of congenital pseudarthrosis. J. Bone & Joint Surg. 31 A: 23–38, 1949.

Murken, ? .: Doppelseitige symmetrische Femurschaftfraktur beim Neugeborenen bei spontaner Geburt. Zbl. f. Gynäk. 51: 2236–2238, 1927.

Müller, G. H. & H. A. Sissons: A case of renal rickets simulating "metaphysal dysostosis". J. Bone & Joint Surg. 32 B: 231–239, 1951.

Nielsen, H.: Forelæsninger over klinisk Endokrinologi og beslægtede Emner III. København, Munksgaard 1941: 261–274.

Nørgaard, F.: Osseous changes in Recklinghausen's neurofibromatosis. Acta Radiologica 18: 460– , 1937.

Ollerenshaw, R.: Congenital defects of the long bones of the lower limb. J. Bone & Joint Surg. 7: 528–552, 1925.

Otten, H.-J.: Intrauterine, symmetrische Spontanfraktur beider Oberschenkel. Zbl. f. Gynäk. 64: 1693–1697, 1940.

Pedersen, H. J.: Blaa scleræ. Ugeskr. f. L. 102: 836, 1940.

Rentoul, J. L.: Fracture in utero. Brit. med. J. 1912, II: 13.

Rivier, Ch. & P. Vulliemin: Présentation d'un cas d'ostéomyélite du nouveau-né. Ann. Pædiatr. 180: 329–332, 1953.

Russell, L. W. & F. A. Chandler: Fibrous dysplasia of bone. J. Bone & Joint Surg. 32 A: 323–337, 1950.

Schönenberg, H. & G. Schallock: Zur Kenntnis der Chondrodysplasia calcificans congenita. Ann. Pædiatr. 180: 129–162, 1953.

Scott, C. R.: Congenital pseudarthrosis of the tibia. Am. J. Roentg. 42: 104–109, 1939.

Seedorff, K. S.: Osteogenesis imperfecta. Disp. Aarhus, 1949.

Smith, R. R.: Intra-uterine fracture. Surg., Gynec. & Obstr. 17: 346–356, 1913.

Snure, H.: Intra-uterine fracture. Radiology 13: 362–365, 1929.

Sperling, M.: Ueber die Actiologie der sogenannten intrauterinen Fracturen an den Extremitäten im besonderen der Unterschenkelknoche. Zschr. f. Geburtsh. u. Gynäk. 24: 225–274, 1892.

Starr, K. W.: Delayed union in fractures. 1947, London, Butterword & Co.

Thannhauser, S. J.: Neurofibromatosis (von Recklinghausen) and osteitis fibrosa cystica localisata et disseminata (von Recklinghausen). Medicine 23: 105–148, 1944.

Trier-Mørch, E.: Chondrodystrophic dwarfs in Denmark. Thesis, Copenhagen 1941, Munksgaard.

Uhlmann, E. & A. Grossmann: Von Recklinghausen's neurofibromatosis with bone manifestations. Ann. Int. Med. 14: 225–241, 1940.

Valls, J., Polak, M. & F. Schajowicz: Fibrous dysplasia of bone, J. Bone & Joint Surg. 32 A: 311, 322, 1950.

Watson-Jones, R.: Fractures and Joint Injuries I. 1946, Livingstone, Edinburgh.

Wilson, Ph. D.: A simple method of two-stage transplantation of the fibula for use in cases of complicated and congenital pseudarthrosis of the tibia. J. Bone & Joint Surg. 23: 639–675, 1941.